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症 例

Congenital Jejunal Stenosis in Adult: A Case Report and Review of Literature

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Introduction

Congenital intrinsic stenosis of the small bowel in adult is rare, although well recognized in newborns as a cause of neonatal intestinal obstruction. Up to the present, about 40 adult cases have been found in medical literature. In these cases, however, the stenotic regions are almost always seen in the duodenum, and is indeed rare in the jejunum. In the past, only one case of jejunal stenosis has been reported by MOORE⁷⁾ in 1981.

In this report we present a second case of congenital jejunal stenosis in an adult with review of literature.

Case Report

A 58-year-old man was admitted to our hospital with complaints of epigastric colicky pain and abdominal fullness. He gave a history of intermittent abdominal pain dating back 40 years and it was sometimes persistent for several days, but he had required no hospitalization. During the past 15 years his attacks occurred two or three times per year, on each occasion, he was diagnosed as having stomach cramps and treated with an antispasmodic agent. This time, however, his symptoms of epigastric pain and flatulence were so severe, and such drugs were not effective, that he was asked to admit. Upper GI series, which had been performed in another institution 3 years prior to admission, showed no remarkable changes. His other medical history included lung tuberculosis 20 years previously.

On physical examination, his pulse rate was 112 per minute and regular, blood pressure was 130/90 mmHg., and no abnormality was detected in the cardiovascular or respiratory systems. The abdomen was distended with rebound tenderness and bowel sounds were hyperactive, however, there were no muscular defense or masses. Rectal examination was negative.

Laboratory examination were within normal limits except that the white blood cell count was

Key words: Congenital intestinal stenosis, Jejunum, Adult case.

索引語: 先天性小腸狭窄症, 空腸発生, 成人例.

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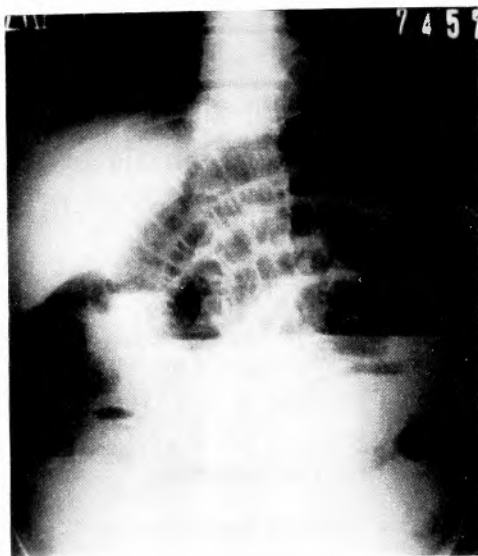


Fig. 1. A plain abdominal roentgenogram showing distended loops of the jejunum on the left upper abdominal quadrant.

17,500/mm³. A plain abdominal roentgenogram showed distended loops of the jejunum on his left upper quadrant (Fig. 1). Obstruction of the small bowel was diagnosed and laparotomy was performed.

During the operation, the small bowel revealed "caliber change" at the jejunum close to the



Fig. 2. Intraoperative photograph showing "caliber change" at the jejuno-ileo transitional region (Arrow).

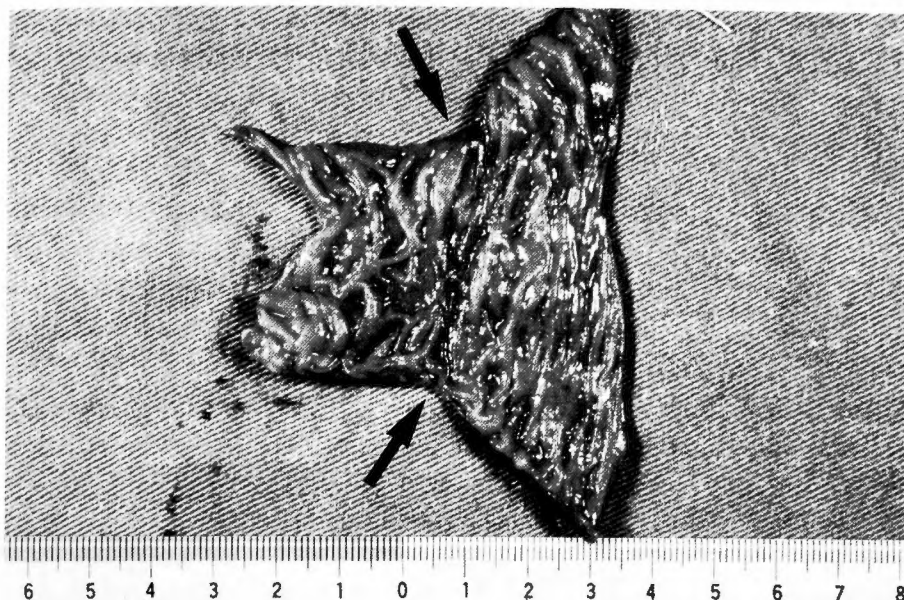


Fig. 3. Resected segment of the jejunum viewed from the mucosal aspect. Arrows point to the stenotic region which did not admit the tip of smallfinger at operation.

jejuno-ileo transitional region and the oral jejunum was massively distended with thick walls, however, there were no evidence of adhesions, strings or bands (Fig. 2). Careful examination of the whole abdominal cavity did not demonstrate any other abnormalities. From these facts



Fig. 4. Microscopic view of the resected specimen showing the thickened muscular layer in the distended jejunum (Hematoxylin and eosin stain, $\times 5$).

an intrinsic abnormality was anticipated, a longitudinal incision at the distended jejunum was made. Jejunotomy demonstrated neither foreign body nor food particles, but revealed a marked stenosis which did not admit the tip of the smallfinger at the site of "caliber change". This region was considered the cause of the obliteration. The small bowel was resected 8 cm in length and end to end anastomosis was done.

His immediate course was uneventful and he was discharged on the 20th postoperative day.

Macroscopic examination of the resected specimen showed similar aspects to congenital jejunal stenosis in a newborn, though there was neither characteristic diaphragm nor windsock. It revealed a streak of mucosal fold which was reddish, circumferential and thickened, and the wall of proximal jejunum to the stenosis was hypertrophic (Fig. 3).

Microscopic examination showed that the stenotic region was composed of normal mucous membrane and submucosa which was slightly folded off, but there was no evidence of mucosal ulceration or fibrosis, and abnormal musculature was not seen. The muscular layer was markedly thickened in the oral distended jejunum, and slight inflammatory cell infiltration was seen in the submucosal layer of the whole resected jejunum. Auerbach's nerve plexus was normal and subserosal layer and serosa revealed nothing in particular (Fig. 4).

Discussion

Congenital intrinsic stenosis or obstruction of the small bowel is often found in newborns, however, is uncommon in adolescents or adults. Since the majority of them reveal clinical symptoms in newborns, the diagnosis has been usually confirmed in the first few days of life. In rare cases, nevertheless, a congenital intestinal stenosis is undetected and remains unnoticed until it causes complete obstruction in adult life.

The first comprehensive case of this disorder in adults was reported by MOORE⁸⁾ in 1884. Since his first report, about 40 adult cases have been reported in world literature^{5,9)}. In these cases, however, the stenotic regions are almost always seen in the duodenum, it is indeed rare in the jejunum or the ileum. Up to the present, only one case in the jejunum has been reported by MOORE et al.⁷⁾ in 1981.

Several theories have been offered to explain the pathogenesis of this disorder. TANDLER's classic theory has been widely accepted since it was written in 1900¹⁰⁾. He states that the lumen of intestine is patent until the fifth week of intra-uterine development when it becomes obliterated by the ingrowth of epithelial cells. A solid cord is formed which normally reopens by vacuolization by the twelfth week. According to this theory, stenosis, atresia or diaphragm is formed when the epithelial cord is not completely resorbed by vacuolization. In this theory, it is characteristic that the stenotic region or diaphragm is composed of a continuation of normal intestinal mucosa and submucosa from an adjacent bowel, and muscle component is not present^{2,9)}. Another plausible theory is an intra-uterine vascular insufficiency from volvulus, internal hernia or intussusception, stated by LOUW et al.⁶⁾ in 1955. The experimental production of intestinal atresia by intra-uterine interruption of the mesenteric blood supply is well known. A complete

disjunction of the intestine or a defect of mesenterium seen in severe cases are satisfactorily explained by this theory¹⁰. The degree of vascular disorder may influence the formation of atresia or stenosis.

In this case, the most likely explanation would seem to relate to intermittent obstruction of the jejunum caused by food particles, which did not result in severe ileus. Therefore, the patient had complained of gastrointestinal symptoms for a long period of time. The marked hypertrophy of the proximal jejunal musculature supports this interpretation. It has also been postulated that there is a relationship between the size of the diaphragmatic opening and the age of the patient at which clinical symptoms are manifested¹¹.

In pathological findings of this case, there was no muscular layer in the partition wall, in addition, inflammatory changes were mild. And there was no defect of mesenterium in the stenotic region. From these facts, a failure of recanalization of intestinal lumen based on Tandler's theory could be considered to explain the possible pathogenesis for this case.

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和文抄録

成人にみられた先天性小腸狭窄症の1例

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先天性小腸狭窄症は，新生児期および乳児期のイレウスの原因として，比較的多くみられる疾患であるが，成人にみられることは非常にまれであり，文献上，現在までに40数例を数えるのみである．しかも成人例においては，その狭窄部位のほとんどが十二指腸であり，空腸や回腸にみられることは極めてまれなケースで，

1981年の Moore の報告があるにすぎない．

今回，われわれは58才の男性で，イレウスの診断のもとに開腹手術を行い，空腸に発生した先天性小腸狭窄症と診断した，世界第二例目と思われる症例を経験したので，その etiology を中心に文献的考察を加え報告した．